## **Aortic Disease Summit**

Hyatt Regency Baltimore on the Inner Harbor September 22–23, 2009

#### AGFNDA



<b>Day I</b> Tuesday, September 22, 2009	Day	Tuesday, September 22,	2009
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8:00	Continental Breakfast
8:30	Opening Remarks  Hal Dietz, MD, Johns Hopkins Univ., Howard Hughes Medical Institute, Baltimore, MD
8:40	GenTAC as an international resource  Kim Eagle, MD, Univ. of Michigan, Ann Arbor, MI

#### Session 1 New Insights into the Pathogenesis of Aortic Aneurysm from Disease Genes and Animal Models

Cheryl Maslen, PhD, Chair 9:00 Developmental underpinnings of apparently acquired aortic aneurysm Hal Dietz, MD, Johns Hopkins Univ., Howard Hughes Medical Institute, Baltimore, MD 9:20 Perturbation of the VSMC contractile unit and aortic aneurysm: clinical presentation, incidence and mechanism Dianna Milewicz, MD, PhD, Univ. of Texas Health Science Center, Houston, TX 9:40 Notch signaling and aortic disease Vidu Garg, MD, Nationwide Children's Hospital, Columbus, OH Angiotensin II induction of thoracic and abdominal experimental aneurysms 10:00 Alan Daugherty, PhD, DSc, Univ. of Kentucky, Lexington, KY Question and Answer 10:20 **Break** 10:35 10:50 Potential Activators of Latent TGFß in Marfan syndrome Daniel Rifkin, PhD, New York Univ., New York, NY AnglI infusion and vascular inflammation through II6 signaling in the mouse model 11:10 Allan Brasier, MD, Univ. of Texas Medical Branch, Galveston, TX

11:30	Phenotype-genotype and genotype-functional correlations in inherited aneurysm conditions Bart Loeys, MD, PhD, Ghent Univ. Hospital, Ghent Belgium
11:50	Transcriptional regulation of coronary artery development  Craig Basson, MD, PhD, Weill Medical College of Cornell Univ., New York, NY
12:10	Question and Answer

## 12:30-1:10 Lunch

# Session 2 Advances in the Imaging and Biomarkers of Aortic Disease

Kathryn Holmes, MD, MPH, and Jonathan Weinsaft, MD, MA, Co-Chairs

1:10	Prognostic utility of assessment of aortic wall biomechanics  João Lima, MD, Johns Hopkins Univ., Baltimore, MD
1:30	Emerging imaging strategies for aortic disease  Andrew Arai, MD, National Institutes of Health, Bethesda, MD
1:50	Biomarker development for assessment of Marfan Syndrome  Jennifer Van Eyk, PhD, Johns Hopkins Univ., Baltimore, MD
2:10	The potential for development of a circulating marker for acute aortic dissection: opportunities and challenges Toru Suzuki, MD, PhD, Univ. of Tokyo, Tokyo, Japan







2:30 Question and Answer

2:50 Break



# Session 3 Gaps in the Natural History of Aortic Disease

Reed Pyeritz, MD, PhD, Chair

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3:10	Incidence and outcome of aortic disease in Turner syndrome  Michael Silberbach, MD, Doernbecher Children's Hospital, Portland, OR
3:25	Aortic dilation in Turner syndrome: progression over time and response to pharmacologic treatment Carolyn Bondy, MD, National Institutes of Health, Bethesda, MD
3:40	Aging and predisposition in Marfan syndrome  Reed Pyeritz, MD, PhD, Univ. of Pennsylvania Medical Center, Philadelphia, PA
4:00	Phenotypic heterogeneity and determinants in aortic aneurysm syndromes  Guillaume Jondeau, MD, Hospital Bichat, Paris, France
4:20	Clinical association of intracranial and aortic aneurysms: a shared genetic basis?  Dong Kim, MD, Univ. of Texas Health Science Center, Houston, TX
4:40	Population prevalence of TAA and its relation to CV risk factors  Richard Devereux, MD, Weill Medical College of Cornell Univ., New York, NY
5:00	Question and Answer
5:30	RECEPTION IN PISCES ON THE 15TH FLOOR

# Day 2 Wednesday, September 23, 2009

**Question and Answer** 

#### 8:00 Continental Breakfast

11:10

#### Session 4 Controversies and Opportunities in the Surgical Management of Aortic Disease

Session 4	Scott LeMaire, MD, and Howard Song, MD, PhD, Co-Chairs
8:30	Surgical considerations regarding the proximal aorta in bicuspid aortic valve syndrome and other non-Marfan entities Joseph Bavaria, MD, Univ. of Pennsylvania Medical School, Philadelphia, PA
8:45	Early outcomes after valve-sparing root replacement: report from the Aortic Valve Operative Outcomes in Marfan Patients Registry Joseph Coselli, MD, Baylor College of Medicine, Houston, TX
9:00	Utility of the Ross procedure in inherited ascending aortic aneurysm  Giovanni Battista Luciani, MD, Univ. of Verona, Verona, Italy
9:20	Current strategies for follow-up and treatment for Ehlers-Danlos syndrome type IV, the "vascular" type Peters Byers, MD, Univ. of Washington, Seattle, WA
9:35	A critical assessment of surgical/interventional practices in vascular EDS  James H. Black, MD, Johns Hopkins Univ., Baltimore, MD
9:50	Question and Answer
10:10	Break
10:30	The role of thoracic endografts in acute aortic syndromes  Lars Svensson, MD, PhD, Cleveland Clinic, Cleveland, OH
10:50	Development of rational surgical practices in patients with TGFB mutations  Duke Cameron, MD, Johns Hopkins School of Medicine, Baltimore, MD







Session 5	Late-breaking science Richard Devereux, MD, Chair
11:20	Deletion of Acta2 Leads to Increased Smooth Muscle Cell Proliferation and Neointimal Formation in Mice  Christina L. Papke Univ. of Texas Health Science Center, Houston, TX
11:30	Analysis of TGFbeta Function in Aortic Aneurysms  Mohamad Azhar, PhD Univ of Arizona, Tucson, AZ
11:40	Copy number variant analysis of patients with sporadic thoracic aortic aneurysms and dissection: Association of the aortic disease with chromosome 16p13.1 duplications encompassing MYH11 gene  Siddharth Prakash, MD, PhD, Baylor College of Medicine, Houston, TX
11:50	Membrane type-1 matrix metalloproteinase plays a multifunctional role in thoracic aortic aneurysm development. Jeffrey A. Jones PhD, Medical University of South Carolina, Charleston, SC
12:00	Smooth muscle cells of fibulin-4 deficient mice contribute to aneurysm progression: An animal model for human thoracic aortic aneurysms  Hiromi Yanagisawa, MD, MPH, Univ. of Texas Southwestern Medical Center, Dallas, TX
12:10	Question and Answer
Session 6	Working Lunch Dianna Milewicz, MD, PhD, and Craig Basson, MD, PhD, Co-Chairs
12:20	GenTAC investigators and others interested get together and reset registry research priorities based on the presentations given. Where do we need to move the science (current and future)?
Session 7	Novel Medical Therapeutic Strategies in the Treatment of Aortic Disease  Hal Dietz, MD, Chair
2:40	AT2 signaling and aortic aneurysm Jennifer Habashi, MD, The Johns Hopkins Hospital, Baltimore, MD
3:00	Novel therapeutic targets in Marfan syndrome  Hal Dietz, MD, Johns Hopkins Univ., Howard Hughes Medical Institute, Baltimore, MD
3:20	Emerging therapeutics to attenuate TGFß signaling  John McPherson, PhD, Genzyme Corporation, Cambridge, MA
3:40	Question and Answer
4:00	Angiotensin receptor blockers: opportunities and limitations  *Robert Thompson, MD, Washington Univ. School of Medicine, St. Louis, MO
4:20	Assessment of infrastructure for clinical trials in vascular disease  Patrice Desvigne-Nickens, MD, National Heart, Lung and Blood Institute, Bethesda, MD
4:40	Question and Answer
5:00	Closing Remarks

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